Source: Finnish Medical Society Duodecim. Systemic Lupus Erythematosus (SLE). Helsinki, Finland: Duodecim Medical Publications Ltd.; 2001 Apr 30.

Systemic lupus erythematosus

EBMG
30.04.2001

Contents

Introduction

- SLE is a syndrome characterized by clinical diversity, changes in the disease activity over time and by aberrant immunological findings.

Epidemiology

- The prevalence of SLE worldwide is 4 - 250 per 100,000. The incidence is most frequent in women aged 15 - 25 years.

Clinical presentation

- The clinical presentation varies between different patients, and in a single patient the disease activity varies over time.
- Constitutional symptoms such as fatigue and fever are common.
- A vast majority of the patients have arthralgia, mostly of the hands.
- About one-half of the patients have cutaneous features, such as malar rash and discoid lupus as well as photosensitivity.
- About one-third of the patients have oral ulcerations.
- About 50% of the patients have nephropathy, which varies from mild proteinuria and microscopical hematuria to end-stage renal failure.

- About 20 - 40% of the patients have pleurisy. Acute pneumonitis and chronic fibrotising alveolitis are relatively rare.
- Pericarditis is somewhat more uncommon than pleuritis. T-wave changes in the ECG are usual.
- Depression and headache are the most common of the neuropsychiatric symptoms. Grand-mal seizures and organic psychoses are rare. A peripheral neuropathy is observed in about 10% of the patients and as many patients get a thromboembolic or hemorrhagic complication of the brain.
- The lymph nodes may enlargen especially when the disease is active.
- There is a risk of first and second trimester foetal losses and of premature birth.

Laboratory findings

- Laboratory findings are diverse.
- Sedimentation rate is usually elevated, the CRPvalue is usually normal.
- Mild or moderate anaemia is common. A clear-cut hemolytic anemia is seen in less than 10% of the patients.
- Leukocytopenia (lymphocytopenia)
- Mild thrombocytopenia
- Antinuclear antibodies are found in over 90% of the patients.
- Anti-DNA antibodies (in 50 - 90% of the patients)
- Polyclonal hypergammablobulinemia
- Decreased complement values (C3 and C4)
- Antiphospholipid antibodies
- Proteinuria, microscopic hematuria, decreased creatinine clearance

Diagnosis

- There is no single symptom or finding that in itself is sufficient for making the diagnosis.
- When SLE is suspected the basic laboratory investigations are:
    - blood count
    - platelets
    - sedimentation rate
    - anti-nuclear antibodies
    - dipstick test of the urine and urinanalysis.
- The diagnosis is based on the clinical symptoms and the laboratory findings and on the ARA classification criteria (1982).
- The patient should be referred to a specialist for evaluation.

Treatment

- The treatment is always individual and depends on the manifestations and activity of the disease. There is no need for treatment solely on the basis of the immunological findings.
- The patients should be encouraged to restrain from sunbathing and to use sunscreens.
- The most important drugs are:
    - nonsteroidal anti-inflammatory drugs
    - hydroxychloroquine (Level of Evidence = C; Evidence Summary available on the EBM Web site)
    - corticosteroids

    - immunosuppressive drugs (e.g. azathioprine, cyclophosphamide)
- Hydroxychloroquine and nonsteroidal anti-inflammatory drugs are used in the treatment of mild symptoms such as cutaneous manifestations and arthralgia. When the response is insufficient or when the patient has fatigue or fever a low dose of corticosteroids (prednisolone 5 - 7.5 mg/day) can be added.
- In the treatment of pleuritis or pericarditis larger amounts of corticosteroids (about 30 mg prednisolone per day) are used.
- In the treatment of severe CNS symptoms and of severe glomerulonephritis, thrombocytopenia and hemolytic anaemia large corticosteroid doses and other immunosuppressive drugs are used (Level of Evidence = A; Evidence Summary available on the EBM Web site).
- The differential diagnosis between an infection and a flare of the SLE is of utmost importance.
- Other drugs that the patient might need, such as antihypertensive treatment, should be remembered.
- If there are signs of renal manifestations the patient should be referred to a nephrologist for a renal biopsy.
- The patients are often allergic to a variety of antibiotics, especially sulfonamides.

Primary antiphospholipid syndrome

- A syndrome manifesting as recurrent venous or arterial thrombotic events, recurrent miscarriages, thrombocytopenia and antiphospholipid antibodies, but without other features of SLE.

Related evidence

- Fluocinonide cream is more effective than hydrocortisone for discoid lupus erythematosus (Level of Evidence = C; Evidence Summary available on the EBM Web site). Hydroxychloroquine and acitretin are as effective.

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